

Zimmermann (6)

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CONGENITAL UNILATERAL ANOPHTHALMUS.

BY

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DISLOCATION OF THE LENS INTO THE ANTERIOR CHAMBER WITH IRIDODIALYSIS; EXTRACTION; RECOVERY.¹

By C. ZIMMERMANN, M.D., MILWAUKEE, WIS.

M. B., a healthy-looking man, fifty-three years old, came to me November 3, 1892, with the statement that at 1 A.M. he had received a severe blow on his left eye with the stick of a whip by a man whom he attempted to push downstairs. A surgeon was sent for at once, who put a stitch through a wound of the upper lid, below the eyebrow, 2 cm long, and referred the case to me in regard to the eye. There was another more superficial wound of the lower lid near and parallel to the ciliary border, 1.50 cm long, which had not been sewed. Both lids were very much swollen and the ocular conjunctiva chemotic. There was no penetrating wound of the globe, but the lower nasal third of the iris was detached from its ciliary border. The pupil was drawn upward and the margin, formed by the separated portion of the iris, was changed into a straight line. The natural and the traumatic pupil looked black; no blood in the anterior chamber. The severed part of the normally blue iris had a green hue and lay farther back than the remaining iris. The cornea showed striated opacities, chiefly in a horizontal direction, the seat of which appeared to be Descemet's membrane. On ophthalmoscopic examination, the artificial as well as the natural pupil gave a red reflex, and in the latter a dark arc with its convexity upwards was noticed, the border of the lens, the lens being dislocated into the anterior chamber. The detached portion of the iris was magnified through the lens, and the green tint seen to be due to the amber color of the latter. V = fingers in 5 feet. V. F. normal. No

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double vision. A pressure bandage was put on. Chemosis and the corneal opacities disappeared in a few days, and the lens became much more clearly visible, looking like a drop of oil in the anterior chamber. The eyeball still red, the upper lid œdematous, but there was no pain at all. Tension not increased. Nov. 18th he complained of pain on the top of his head and at the infra-orbital margin. Nov. 19th he slept well, but the œdema of upper lid not disappearing and the eyeball being redder with increase of intraocular pressure, I advised extraction of lens. The patient consenting, I removed the lens Nov. 20th under aseptic measures, with von Graefe's knife, by upper corneal section, the apex of it 2 mm below the limbus. Although vitreous prolapsed as soon as the incision began, I quietly finished it. The speculum was removed at once and the lens expelled within its capsule by gently pressing the lower lid with my left index finger on the lower part of the eyeball towards its centre, while I directed the upper lid and the peripheral edge of the wound with my right forefinger. The patient, being very unruly (despite cocaine) lost some vitreous. After he was bandaged he complained of pain in the eye, which however was promptly relieved by antipyrine. Nov. 24th the first change of dressing (4th day). No discharge, wound nicely closed, no prolapse of iris. At the temporal corner of the wound a small gray thread of vitreous projected, which I left undisturbed, lest by snipping it off the wound might burst, until Dec. 27th, when I cut it off. Feb. 9th V with $+ 10 D = \frac{15}{200}$.

Details of the fundus can be seen, although a little veiled by slight opacities of the vitreous, appearing in horizontal wave-like dark lines. The eye is without the least irritation. The detached portion of the iris has now the same color as the remainder. This piece being in the centre, it impairs the sight to a certain extent. The patient however mostly uses his artificial pupil and is not annoyed by monocular double images. So far I thought it to be too great a risk to cut this piece with intraocular scissors, as not much could be gained.

Some features in this case deserve especial attention:

1. The manner in which the injury happened is quite interesting. The blow must have struck the upper portion of the eyeball with such vehemence, that the upper periphery of the lens was pushed backwards and turning on

its transverse diameter as axis was driven with its lower periphery against the iris, tearing the latter from its ciliary margin and entering through this gap into the anterior chamber, so that the posterior surface of the lens was now in front and towards the cornea, the lower lens border turned upwards.

2. The great force of the blow explains also the absence of hemorrhages from the ruptured blood-vessels of the iris.

3. In regard to the corneal opacities seen at first, the objection might be made, that I erroneously located them in the cornea, whereas they were in reality in the lens touching the cornea, and were analogous to the apparent opacity of the lens, which may be observed, when after sudden advancement of the latter by evacuation of the aqueous, the layers of the lens suffer a displacement along each other within the capsule and reflect incident light like a soft cataract (*cf.* Becker in *Graefe-Saemisch*, v., p. 280). But then they would not have disappeared as they did after a few days, as the lens did not change its position next to the cornea. In my opinion they represented the optical appearance of wrinkles of Descemet's membrane and the deeper corneal layers, in a horizontal direction, caused by a compression and flattening of the eyeball from above downwards, in consequence of the blow acting from above. Thus the horizontal meridian of the cornea was stretched and its radius elongated, that of the vertical meridian shortened. Hess (*von Graefe's Arch.*, xxxviii., 4, p. 1) proved anatomically and experimentally, that the striated opacities of the cornea after cataract extractions are to be explained in the same way (the direction of the compression only differing from that in our case). They consist in wave-like wrinkles of the deeper corneal strata and are produced by the difference in curvature of the vertical and horizontal meridians of the cornea, the radius of the vertical meridian becoming larger after the opening of the eyeball, acting as if the eyeball was compressed from both sides. In our case the opacities disappeared very soon, the eyeball gradually reassuming its natural curvature, thus furnishing a further support of this explanation.

CONGENITAL UNILATERAL ANOPHTHALMUS.

BY C. ZIMMERMANN, M.D., MILWAUKEE, WIS.

CONGENITAL anophthalmus is mostly bilateral and has been found to occur in different degrees, either limited to the eyeball, or to the eyeball and the optic nerve and its central terminations, or even associated with defect of a large area of the brain. The adnexa of the eyeball are mostly developed. Unilateral anophthalmus, called also monophthalmus, is much rarer. (It must not be confounded with cyclopia, which means confluence of both eyes into one double eye in an abnormal place, viz., at the region of the glabella.) The time of its development is attributed to a late period of foetal life, if only the globe is missing, not its appendages. In most cases remnants of a globe were left, sometimes not as large as a pea, characterized as such by the presence of choroidal pigment. Others showed orbital cysts, starting from such rudiments, and in others no trace of an eyeball could be detected. The theory is, that by external causes the eyeball at first well developed suffered an arrest of evolution, and consequently degeneration and complete destruction. Nearly all observations were made in children. I saw a case in an adult, which, as it exhibited some other malformations besides, may be of sufficient interest for a short communication.

The patient, a robust man, thirty-three years old, six feet one inch high, weighing two hundred and twenty-seven pounds, is perfectly intelligent. He is the third child of healthy parents

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(not consanguineous), and neither these nor his sisters and brothers suffered from any malformations. The right eye is perfectly healthy and normally shaped in all its portions and appendages, V., V. F., and ophthalmoscopic condition normal. The left eyeball is wanting. Its adnexa are in their normal place and all developed, but of diminished size. The lids are smaller, the upper one slightly entropic with its ciliary margin touching the lower one, which is a little ectropic. The palpebral fissure is 2 *cm* long (in right eye 3.5 *cm*). Caruncle, lachrymal points, and lachrymal gland are there, tears flowing from the eye just as from its fellow on irritation. After insertion of lid-retractors a diminished empty conjunctival sac is exposed, in the depth of which no trace of an eyeball can be seen nor felt. The finger finds a soft cushion, feeling as the contents of the orbit do after enucleation. The conjunctiva at the bottom of the sac looks a little whitish from the underlying Tenon's capsule. The presence of the ocular muscles becomes evident by their action, as the sac moves in all directions in association with the other eye. Neither pressure nor the faradic current creates any sensation of light, so that absence or degeneration of the optic nerve may be inferred. The left orbit is considerably smaller than the right. The height at the supra-orbital foramen : L, 2 *cm* ; R, 3 *cm*. Greatest horizontal width : L, 3 *cm* ; R, 4 *cm*. Left eyebrow 5 *cm* long ; R, 6.5 *cm*.

The skull is asymmetric, the left portion of face and forehead being smaller than the right. The left side of the forehead is receding as well as the hair on this side. The following measures were taken with the 'compass : Longitudinal diameter (root of nose to occipital protuberance), 18 *cm*. Root of nose to tuberculum radialis arcus zygomatici (above and behind the ear) L, 12 *cm* ; R, 12.5 *cm*. The width between both ears, *i. e.*, between the above-mentioned tubercula, 15 *cm*. Width between both tubera parietalia, 16 *cm*. Distance of external walls of both orbits, 10 *cm*. Figures obtained with tape-measure : Horizontal circumference of head, taken from centre between both arcus superciliares to occipital protuberance, 56 *cm*. The left (pathological) half of it measures 29 *cm*, the right only 27 *cm*, the left parietal bone showing a greater extension than the right, compensating for the diminished size of the left side of the forehead. Root of nose to the external wall of left orbit, 6 *cm* ; of right orbit, 7 *cm*. Frontal arch between the roots of both arcus zygomatici, 32.5 *cm*. The

patient has a cranium progenæum, the lower jaw forming an overbite over the superior maxillary. The palatal vault is very high and narrow. The left superior maxillary is smaller than the right and has one tooth, the canine, less than the right. The posterior wall of the pharynx projects more on the right than on the left side. In posterior rhinoscopy the left choana appears to be smaller than the right. The deviation of the nasal septum to the right in front is due to a fracture he suffered when fifteen years old.

The left auricle shows also a deformity. Its lobe is rudimentary, and the whole auricle looks as if it had been rolled up in a forward direction and drawn forward. Its posterior aspect nearly forms a plane, without bends and elevations, and stands out at right angle with the head, so that the anterior limit of the helix lies in one frontal plane with the posterior wall of the external ear opening. In a profile view it seems to cover half of the concha. In the skin, 0.75 *cm* in front of the tragus, is an elevated scar 1 *cm* long (vertical diameter) and 0.56 *cm* wide (sagittal diameter) where, according to the patient's statement, the auricle was attached at birth and was severed by operation soon afterwards. A small scar at the posterior surface of the helix corresponds exactly to that place. When the auricle is rolled forward, both scars touch each other. After the inspissated cerumen was removed, the auditory canal and *Mt* appear perfectly normal, as well as the hearing power, so that the malformation of the hearing organ is limited to the auricle. There is no other asymmetry, except that the left foot is thicker than the right.

The story the parents give in this case is this: The boy was born in a position in which he held his left hand on the left eye, pulling the left auricle forward with his fingers. The eye was wanting, and the ear was firmly attached to the head. The left leg was drawn upward and, when stretched, tended to return to that position, so that it had to be fixed on splints. Hæderath (Ueber Monophthalmus congenitus, Inaug. Diss., Bonn, 1871) described two similar cases observed in the eye-clinic at Bonn, one of which is just like ours, if not the same, since our patient has been examined by Professor Sæmisch of Bonn in 1867, when seven years old. Hæderath, however, reports his age as eleven years

and gives a different first initial. He writes, that the left arm retained the same position on the eye as at birth for a fortnight afterwards. Manz (*Græfe-Sæmisch*, ii., p. 125), in quoting this case of H., seems to consider the anophthalmus to be of traumatic origin, caused by the pressure of the hand, leaving it undecided whether the peculiar position of the arm may have been due to a constriction by amniotic threads, or the umbilical cord. If this had been the case, the constriction must have been very firm, so that we would expect to find some traces of it on the arm or hand, an atrophic spot or a scar, or some other malformation, but there was nothing of the kind. As not only the eye and ear, which alone might have suffered from the pressure of the hand, but even the whole left side of the head were defective, this factor loses its value as the only cause of the malformation. The child's condition at birth not having been communicated by an expert, we are not allowed to adopt at once the parents' pretty vague narration and base theories on it. The father told me that at first they thought the midwife had injured the eye and had invented that story for her exculpation. Apparently he had not seen the asserted position, nor had the mother, as they at first tried to conceal from her the existence of such a deformity. The case however is of especial interest, as being one of those rare instances in which the anophthalmus was observed in an adult, and corroborating the assumption that this malformation was due to an arrest of development, with consequent destruction of the eyeball in a pretty late period of foetal life, when the eyeball was already developed, as we infer from the presence of its appendages, and not caused by a primary deficiency; else we could not expect such perfectly healthy mental and otherwise bodily development as seen in our patient.

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